9.7.3. Infectious Ocular Inflammatory Diseases (III): HSV and VZV (III): Acute Retinal Necrosis

diagnostic tests

- polymerase chain reaction (PCR) testing of aqueous and/or vitreous samples
- retinal choroidal biopsy
- Intraocular antibody production
  - Goldmann-Witmer (GW) coefficient
    - aqueous humor analysis
      - anterior chamber paracentesis
    - ratio >3 is diagnostic of local antibody production
    - adjunct to the diagnosis of toxoplasmosis and uveitis caused by HSV and VZV infection
    - little value in the diagnosis of CMV retinitis
    - combining the GW coefficient with PCR analysis may increase diagnostic yield
  - PCR testing
    - most sensitive, specific, and rapid diagnostic method for detecting infectious posterior uveitis in general and ARN specifically
    - has largely supplanted viral culture, intraocular antibody titers, and serology
    - aqueous humor
      - aqueous sampling is usually sufficient
    - vitreous
      - quantitative PCR
        - information with respect to viral load, disease activity, and response to therapy
      - etiology
        - HSV-1 or VZV
          - tend to be older (mean age, 40 years)
        - VZV> HSV-1> HSV-2
          - HSV-1
            - higher risk of encephalitis and meningitis
          - HSV-2
            - younger (<age 25 years)
        - in rare instances, CMV
- endoretinal biopsy
  - rarely when PCR results are negative but the clinical suspicion is high

treatment

- induction antiviral therapy
  - Intravenous acyclovir, 10 mg/kg every 8 hours for 10–14 days,
    - effective against HSV and VZV
    - ← reversible elevations in serum creatinine and liver enzymes
    - in the presence of frank renal insufficiency, the dosage will need to be reduced
  - oral valacyclovir at doses of up to 2 g 3x/day
    - alternative to intravenous acyclovir
    - has not been demonstrated as superior to the intravenous approach
  - intravitreal antiviral
    - ganciclovir (0.2–2.0 mg/0.1 mL)
    - foscarnet (1.2–2.4 mg/0.1 mL)
    - for rapid induction (in combination with intravenous or oral antivirals)
    - for disease that fails to respond to systemic acyclovir
    - injections may need to be repeated twice weekly until the retinitis is controlled
  - treatment prevents development of new lesions and promotes lesion regression over 4 days
- systemic corticosteroids
  - oral prednisone, 1 mg/kg/day after 24–48 hours of antiviral therapy
  - tapered over several weeks
- aspirin and other anticoagulants
  - to prevent vascular occlusions
  - results have been inconclusive
- maintenance antiviral therapy
  - extended antiviral therapy may reduce the incidence of contralateral disease by 80% over 1 year
    - x3 months
  - VZV infection
    - acyclovir at 800 mg orally 5 times daily, valacyclovir at 1 g orally 3 times daily, or famciclovir at 500 mg orally 3 times daily
  - HSV-1
    - dose is one-half of that for VZV
- retinal detachment
  - prophylactic
    - prophylactic barrier laser photocoagulation
      - as soon as view permits
      - to areas of healthy retina at posterior border of necrotic lesions
      - may prevent retinal detachment
    - early vitrectomy combined with endolaser photocoagulation
      - to eliminate the contribution of vitreous traction on the necrotic retina
  - vitrectomy, air–fluid exchange, endolaser photocoagulation, long-acting gas or silicone oil tamponade
    - more successful than standard scleral buckling
- prognosis
  - untreated
    - final visual acuity of ≤20/200 in 2/3
  - with early recognition, aggressive antiviral therapy, and laser photocoagulation, this prognosis may be significantly improved
    - 46% achieved a visual acuity of ≥20/40
    - 92% achieved a visual acuity of ≥20/400

necrotizing herpetic retinopathy

- retinal lesions of presumed herpetic etiology that are not characteristic of well-recognized syndromes such as ARN, CMV retinitis, or progressive outer retinal necrosis

Acute retinal necrosis (ARN)

- epidemiology
  - otherwise healthy adults
  - immunocompromised patients (including HIV infection)
  - M=F
  - clustering in fifth-seventh decades
    - reported in children
- prodromal infection
  1. without a systemic prodrome years after primary infection
  2. following cutaneous herpetic infection
     - dermatomal zoster
  3. following systemic herpetic infection
     - chickenpox
     - herpetic encephalitis
       - HSV-1 or -2

diagnostic criteria

- See Table 7-1

clinical presentation

- symptoms
  - loss of vision
  - photophobia
  - floaters
  - pain
  - acute unilateral
- fellow eye involvement
  - 36%
  - usually within 6 weeks
    - may be delayed for extended periods
- panuveitis
  - significant anterior segment inflammation
    - KPs
    - posterior synechiae
    - elevated IOP
  - heavy vitreous cellular infiltration
- within 2 weeks
  - classic triad
    1. occlusive retinal arteriolitis
    2. vitritis
    3. multifocal yellow-white peripheral retinitis
       - early
         - peripheral retinal lesions are discontinuous and have scalloped edges
         - appear to arise in the outer retina

Figure 7-3 (© 2020 American Academy of Ophthalmology)

       - within days
         - lesions coalesce to form a confluent 360° creamy retinitis
         - progresses in a posterior direction
         - full-thickness retinal necrosis
         - occasional retinal hemorrhage
         - posterior pole tends to be spared

Figure 7-4 (© 2020 American Academy of Ophthalmology)

- combined tractional–rhegmatogenous retinal detachments
  - 75%
  - multiple posterior retinal breaks
  - within the first weeks to months
  - proliferative vitreoretinopathy

Figure 7-5 (© 2020 American Academy of Ophthalmology)

- exudative retinal detachment
- disc swelling and relative afferent pupillary defect

differential diagnosis

- CMV retinitis
- atypical toxoplasmic retinochoroiditis
- syphilis
- lymphoma
- leukemia
- autoimmune retinal vasculitis (such as Behçet disease)

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